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Case Report

# A Case of Super Refractory Status Epileptics (SRSE)

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## Abstract

Super-refractory status epilepticus (SRSE) is defined as status epilepticus that continues or recurs 24 h or more after the onset of anaesthetic therapy, including those cases where status epilepticus recurs on the reduction or withdrawal of anaesthesia.

The condition may be triggered by as yet unidentified infections or an immunological mechanism and is associated with high mortality and morbidity rates. However, an early, aggressive and meticulous supportive care results in good outcomes.

We present a case report of a 31year old female, who reported with a prodrome of headache and vomiting to our Tertiary care hospital. She developed refractory multifocal and generalised seizures followed by altered sensorium (GCS- E2V1M5), which required intensive care in ICU.

She was managed with aggressive and proactive Intensive care by early Intubation (Day1) and mechanical ventilation support. She was initially treated with Anti Epileptic drugs- intravenous phenytoin sodium, Sodium valproate and levetiracetam, antivirals and antibacterials. Later IV steroids and IV immunoglobulin were added. Burst suppression was achieved with iv anaesthetic (Thiopental). Good meticulous supportive care enabled an early weaning off the ventilator by day 18 and we were able to discharge the patient out of hospital by Day 30 with good recovery.

***Keywords:*** *SRSE; Drugs; Treatment*

## Introduction

Super-refractory status epilepticus (SRSE) is defined as status epilepticus that continues or recurs 24 h or more after the onset of anaesthetic therapy, including those cases where status epilepticus recurs on the reduction or withdrawal of anaesthesia.

The condition may be triggered by as yet unidentified infections or an immunological mechanism. The term new onset refractory status epilepticus (NORSE) syndrome has been used to describe patients who develop refractory status epilepticus with no prior history of epilepsy, or identifiable causative factors [1,2]. It is an uncommon clinical problem with high mortality and morbidity rates; However, an early, aggressive and meticulous supportive care has been associated with good outcomes [3,4].

## Case Report

A 31year old female reported with a prodrome of headache and vomiting.

She developed refractory multifocal and generalised seizures followed by altered sensorium (GCSE2V1M5), which required admission to the intensive care unit. She received anti epileptic drugs (AED) treatment with intravenous phenytoin sodium, Sodium valproate and levetiracetam but developed refractory status epilepticus requiring induction of general anaesthesia with intravenous Thiopentone at high doses of 10mg/kg/hr and protection of airway with ETT and mechanical ventilation.

Seizure control was monitored clinically as well as using the cerebral function monitor (CFAM) and intermittent electroencephalogram (EEG) recording. She developed a moderate grade fever and had mild leucocytosis. Her MR imaging, CSF examination and serological tests for infective and autoimmune aetiologies were normal.

She was initially treated with broad spectrum antibacterials and antiviral (acyclovir). IV steroids were administered on day 02 (Methyl prednisolone 1g for 3 days and then oral prednisolone 60 mg/day) and IV immunoglobulin 150 g over 5 days from day 07 onwards. Along with these she was put on four AEDs (Topiramate, Leviteracetam, Phenytoin sodium and Lacusonamide). She was placed on continuous EEG monitoring and total burst suppression was achieved with no activity for >10 sec for next 84-90 hours.

Barbiturate induced hypotension was managed by dopamine infusion and advanced haemodynamic monitoring. Seizures recurred on withdrawal of barbiturate anaesthetic until day 15 for which iv Thiopentone was reinstituted until total suppression of seizure activity achieved.

As a part of intensive and aggressive care an early Intubation (Day1) was done for protection of airway and tracheostomy done on Day 10 of her stay in ICU. Regular suction and chest physiotherapy was given and she was weaned off the ventilator on Day 18 and tracheostomy was closed by Day 25. She developed Critical illness neuropathy which was taken care of by iv pyridoxine, prosthetic splints and physiotherapy. Occasional seizures reoccurred on withdrawal or tapering of AEDs and steroid therapy, which was acceptable and was put on long term AE treatment with 4 AEDs. She made a good recovery by the time of discharge.

## Discussion

Tonic–clonic status epilepticus is a medical emergency. Treatment is aimed at stopping seizures largely in order to avoid cerebral damage and other morbidity.

Typically, in Stage 1 (early status epilepticus), therapy is with benzodiazepines.

If seizures continue despite this therapy, the patient is said to be in Stage 2 (established status epilepticus) and therapy is with intravenous anti-epileptic drugs such as phenytoin, phenobarbital or valproate.

If seizures continue despite this treatment for up to 2 h, the patient is said to be in Stage 3 (refractory status epilepticus) and general anaesthesia is usually recommended, at a dose that results in EEG burst suppression (a level of anaesthesia at which all seizure activity is usually controlled).

It is interesting in passing to note that anaesthesia has been recommended since the mid-19th century, and John Hughlings Jackson for instance writes that 'chloral is the best drug; and if the fits are very frequent, etherisation will help' (Hughlings Jackson, 1888) [1].

The status epilepticus is conventionally treated with the full panoply of intensive treatment unit care, including assisted ventilation and full cardiovascular monitoring. Benzodiazepine and barbiturate anaesthetic drugs invariably cause hypotension and cardiorespiratory depression, which is sometimes severe and limits treatment, and pressor agents are usually necessary. Prolonged refractory status epilepticus leads to high mortality. Most patients die from systemic complications more related to their management in the intensive care unit than to brain damage. The need for mechanical ventilation, vasopressors, third line epileptic drugs or inhalational anesthetic agents

lead to hypotension, which is associated with poor outcome [4]. Good care of comatose and intubated patient with regular closed suctioning and chest physiotherapy prevents Ventilator associated pneumonia and other complications [5-8].

## Conclusion

Super-refractory status epilepticus is a serious condition. The mortality rate is substantial, where patients die from systemic complications linked to their ICU stay, or treatment withdrawal rather than to brain damage. Multidisciplinary collaboration is required to establish the diagnosis and appropriate management of patients presenting with SRSE.

In our case, aggressive and meticulous supportive care, with an early mechanical ventilator support, advanced haemodynamic monitoring, early induction of iv anaesthesia and timely weaning, helped patient to walk out of the ICU and hospital with satisfactory outcome.

Since small number of case reports have been published in available literature so far, this case is presented to increase the database of outcomes of individual therapies, in order to establish the treatment and critical care protocol.

### Key messages

- Prolonged refractory status epilepticus leads to high mortality.
- Most patients die from systemic complications more related to their management in the intensive care unit than to brain damage.
- An early, aggressive and proactive Critical care has been associated with good outcome.

### Critical care and anaesthetic goals

- Protection of Airway – Low threshold for Intubation and ventilatory support.
- Neuroprotection with Advanced Haemodynamic monitoring
- Prevention of associated complications (VAP) – Early weaning
- Prevention of critically ill neuropathy
- Limb physiotherapy and Splints.

### Consent

Written informed consent was obtained from the NOK of patient for publication of this manuscript.

## Competing Interests

The authors declare that they have no competing interests. Authors' contribution SS and NSL performed the clinical examination, analyzed and interpreted the diagnostic findings. Conception and discussion was performed by all authors.

The main writing was done by SS and AG.

All authors read and approved the final manuscript.

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